



Complete Summary

GUIDELINE TITLE

Guideline on oral health care/dental management of heritable dental developmental anomalies.

BIBLIOGRAPHIC SOURCE(S)

American Academy of Pediatric Dentistry Council on Clinical Affairs. Guideline on oral health care/dental management of heritable dental development anomalies. *Pediatr Dent* 2008-2009;30(7 Suppl):196-201. [52 references] [PubMed](#)

GUIDELINE STATUS

This is the current release of the guideline.

COMPLETE SUMMARY CONTENT

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SCOPE

DISEASE/CONDITION(S)

Heritable dental developmental anomalies:

- Amelogenesis imperfecta (AI)
- Dentinogenesis imperfecta (DI)
- Dentin dysplasia (DD)

GUIDELINE CATEGORY

Diagnosis
Management
Treatment

CLINICAL SPECIALTY

Dentistry
Pediatrics

INTENDED USERS

Dentists

GUIDELINE OBJECTIVE(S)

To address the diagnosis, principles of management, and objectives of therapy of children with heritable dental developmental anomalies rather than provide specific treatment recommendations

TARGET POPULATION

Children with heritable dental developmental anomalies

INTERVENTIONS AND PRACTICES CONSIDERED

Management of Amelogenesis Imperfecta (AI)

1. Differential diagnosis
2. General considerations and principles of management
 - Comprehensive and timely approach
 - Addressing clinical and emotional demands of patients with sensitivity
3. Preventive care
 - Regular periodic examinations
 - Meticulous oral hygiene
 - Fluoride applications
 - Desensitizing agents
4. Restorative care
 - Bleaching and/or microabrasion
 - Composite resin or porcelain veneers
 - Full coverage restoration
 - Implants or prosthetics
 - Multidiscipline approach

Management of Dentinogenesis Imperfecta (DI)

1. Differential diagnosis
2. General considerations and principles of management
 - Preventing severe attrition associated with enamel loss and rapid wear of the poorly mineralized dentin
 - Rehabilitating dentitions that have undergone severe wear
 - Optimizing esthetics
 - Preventing caries and periodontal disease
3. Preventive care
 - Regular periodic examinations
 - Meticulous oral hygiene

- Calculus removal
- Oral rinses, fluoride applications and desensitizing agents
- 4. Restorative care
 - Routine restoration for mild cases
 - Full coverage restorations in both the primary and permanent dentitions for severe cases
 - Bleaching or use of veneers
- 5. Endodontic considerations
 - Periodic radiographic surveys
 - Apical surgery for abscesses
- 6. Evaluation and multidisciplinary management of malocclusion

Management of Dentin Dysplasia (DD)

1. Differential diagnosis
2. General considerations and principles of management
3. Preventive care
 - Meticulous oral hygiene
4. Restorative care
 - Prosthetic replacement including dentures, overdentures, partial dentures, and/or dental implants
5. Endodontic considerations
 - Periapical curettage and retrograde amalgam seals

MAJOR OUTCOMES CONSIDERED

- Success rate of restoration and other treatments
- Change in emotional/psychological impact of conditions after treatment
- Time to loss of teeth
- Rate of tooth wear and tooth loss

METHODOLOGY

METHODS USED TO COLLECT/SELECT EVIDENCE

Searches of Electronic Databases

DESCRIPTION OF METHODS USED TO COLLECT/SELECT THE EVIDENCE

A MEDLINE search was conducted using the terms "amelogenesis imperfecta", "dentinogenesis imperfecta", "dental dysplasia", "dentin dysplasia", "enamel hypoplasia", "enamel hypocalcification", "amelogenin", and "enamelin".

NUMBER OF SOURCE DOCUMENTS

Not stated

METHODS USED TO ASSESS THE QUALITY AND STRENGTH OF THE EVIDENCE

Not stated

RATING SCHEME FOR THE STRENGTH OF THE EVIDENCE

Not applicable

METHODS USED TO ANALYZE THE EVIDENCE

Review

DESCRIPTION OF THE METHODS USED TO ANALYZE THE EVIDENCE

Not stated

METHODS USED TO FORMULATE THE RECOMMENDATIONS

Expert Consensus

DESCRIPTION OF METHODS USED TO FORMULATE THE RECOMMENDATIONS

Clinical guidelines of the American Academy of Pediatric Dentistry (AAPD) are developed under the direction of the Board of Trustees, utilizing the resources and expertise of its membership operating through the Council on Clinical Affairs (CCA).

Proposals to develop or modify guidelines may originate from 4 sources:

1. The officers or trustees acting at any meeting of the Board of Trustees
2. A council, committee, or task force in its report to the Board of Trustees
3. Any member of the AAPD acting through the Reference Committee hearing of the General Assembly at the Annual Session
4. Officers, trustees, council and committee chairs, or other participants at the AAPD's Annual Strategic Planning Session

Regardless of the source, proposals are considered carefully, and those deemed sufficiently meritorious by a majority vote of the Board of Trustees are referred to the CCA for development or review/revision.

Once a charge (directive from the Board of Trustees) for development or review/revision of a clinical guideline is sent to the CCA, it is assigned to 1 or more members of the CCA for completion. CCA members are instructed to follow the specified format for a guideline. All clinical guidelines are based on 2 sources of evidence: (1) the scientific literature; and (2) experts in the field. Members may call upon any expert as a consultant to the council to provide expert opinion. The Council on Scientific Affairs provides input as to the scientific validity of a guideline.

The CCA meets on an interim basis (midwinter) to discuss proposed clinical guidelines. Each new or reviewed/revised guideline is reviewed, discussed, and confirmed by the entire council.

RATING SCHEME FOR THE STRENGTH OF THE RECOMMENDATIONS

Not applicable

COST ANALYSIS

A formal cost analysis was not performed and published cost analyses were not reviewed.

METHOD OF GUIDELINE VALIDATION

Peer Review

DESCRIPTION OF METHOD OF GUIDELINE VALIDATION

Once developed by the Council on Clinical Affairs (CCA), the proposed guideline is submitted for the consideration of the Board of Trustees. While the board may request revision, in which case it is returned to the council for modification, once accepted by majority vote of the board, it is referred for Reference Committee hearing at the upcoming Annual Session. At the Reference Committee hearing, the membership may provide comment or suggestion for alteration of the document before presentation to the General Assembly. The final document then is presented for ratification by a majority vote of the membership present and voting at the General Assembly. If accepted by the General Assembly, either as proposed or as amended by that body, the document then becomes the official American Academy of Pediatric Dentistry (AAPD) clinical guideline for publication in the AAPD's Reference Manual and on the AAPD's Web site.

RECOMMENDATIONS

MAJOR RECOMMENDATIONS

Amelogenesis Imperfecta (AI)

General Considerations and Principles of Management

A primary goal for treatment is to address each concern as it presents but with an overall comprehensive plan that outlines anticipated future treatment needs. Clinicians treating children and adolescents with AI must address the clinical and emotional demands of these disorders with sensitivity. It is important to establish good rapport with the child and family early. Timely intervention is critical to spare the patient from the psychosocial consequences of these potentially disfiguring conditions. A comprehensive and timely approach is reassuring to the patient and family and may help decrease their anxiety.

Preventive Care

Early identification and preventive interventions are critical for infants and children with AI in order to avoid the negative social and functional consequences of the disorder. Regular periodic examinations can identify teeth needing care as

they erupt. Meticulous oral hygiene, calculus removal, and oral rinses can improve periodontal health. Fluoride applications and desensitizing agents may diminish tooth sensitivity.

Restorative Care

The appearance, quality, and amount of affected enamel and dentin will dictate the type of restorations necessary to achieve esthetic, masticatory, and functional health. When the enamel is intact but discolored, bleaching and/or microabrasion may be used to enhance the appearance. If the enamel is hypocalcified, composite resin or porcelain veneers may be able to be retained with bonding. If the enamel or dentin cannot be bonded, full coverage restorations will be required. In order to facilitate veneer or crown placement, periodontal therapy may be necessary when acute/chronic marginal gingivitis along with hyperplastic tissue exists.

During the primary dentition, it is important to restore the teeth for adequate function and to maintain adequate arch parameters. Primary teeth may require composite or veneered anterior crowns with posterior full coverage steel or veneered crowns.

The permanent dentition usually involves a complex treatment plan with specialists from multiple disciplines. Periodontics, endodontics, and orthodontics may be necessary and treatment could include orthognathic surgery. The prosthetic treatment may require veneers, full coverage crowns, implants, and fixed or removable prostheses. The fabrication of an occlusal splint is advocated to reestablish vertical dimension when full mouth rehabilitation is necessary. Therapy will need to be planned carefully in phases as teeth erupt and the need arises.

Behavior guidance, as well as the psychological health of the patient, will need to be addressed in each phase. Counseling for the child or adolescent and his/her family should be recommended when negative psychosocial consequences of the disorder are recognized. Due to extensive treatment needs, a patient may require sedation or general anesthesia for restorative care.

Dentinogenesis Imperfecta (DI)

General Considerations and Principles of Management

Providing optimal oral health treatment for DI frequently includes preventing severe attrition associated with enamel loss and rapid wear of the poorly mineralized dentin, rehabilitating dentitions that have undergone severe wear, optimizing esthetics, and preventing caries and periodontal disease. The dental approach for managing DI will vary depending on the severity of the clinical expression.

The clinician must be cautious in treating individuals with DI if performing surgical procedures or other treatment that could transmit forces to the jaws, increasing the risk of bone fracture. Some types of protective stabilization may be contraindicated in the patients with DI.

Preventive Care

Early identification and preventive interventions are critical for individuals with DI in order to avoid the negative social and functional consequences of the disorder. Regular periodic examinations can identify teeth needing care as they erupt. Meticulous oral hygiene, calculus removal, and oral rinses can improve periodontal health. Fluoride applications and desensitizing agents may diminish tooth sensitivity.

Restorative Care

Routine restorative techniques often can be used effectively to treat mild to moderate DI. These treatments more commonly are applied to the permanent teeth, as the permanent dentition frequently is less severely affected than the primary dentition. In more severe cases with significant enamel fracturing and rapid dental wear, the treatment of choice is full coverage restorations in both the primary and permanent dentitions. The success of full coverage is greatest in teeth with crowns and roots that exhibit close to a normal shape and size, minimizing the risk of cervical fracture.

Ideally, restorative stabilization of the dentition will be completed before excessive wear and loss of vertical dimension occur. Cases with significant loss of vertical dimension will benefit from reestablishing a more normal vertical dimension during dental rehabilitation. Cases having severe loss of coronal tooth structure and vertical dimension maybe considered candidates for overdenture therapy. Overlay dentures placed on teeth that are covered with fluoride-releasing glass ionomer cement have been used with success.

Bleaching has been reported to lighten the color of DI teeth with some success; however, because the discoloration is caused primarily by the underlying yellow-brown dentin, bleaching alone is unlikely to produce normal appearance in cases of significant discoloration. Different types of veneers can be used to improve the esthetics and mask the opalescent blue-gray discoloration of the anterior teeth.

Endodontic Considerations

Some patients with dentinogenesis imperfecta will suffer from multiple periapical abscesses apparently resulting from pulpal strangulation secondary to pulpal obliteration or from pulp exposure due to extensive coronal wear. The potential for periapical abscesses is an indication for periodic radiographic surveys on individuals with DI. Because of pulpal obliteration, apical surgery may be required to maintain the abscessed teeth. Attempting to negotiate and instrument obliterated canals in DI teeth can result in lateral perforation due to the poorly mineralized dentin.

Occlusion

Class III malocclusion with high incidences of posterior crossbites and openbites occur in DI Type I and should be evaluated. Multidisciplinary approaches are essential in addressing the complex needs of the individuals affected with DI.

Dentin Dysplasia (DD)

General Considerations and Principles of Management

The goal of treatment is to retain the teeth for as long as possible. However, due to shortened roots and periapical lesions, the prognosis for prolonged tooth retention is poor. Prosthetic replacement including dentures, overdentures, partial dentures, and/or dental implants may be required.

Preventive Care

Preventive care is of foremost importance. Meticulous oral hygiene must be established and maintained. As a result of shortened roots with DD Type I, early tooth loss from periodontitis is frequent.

Restorative Care

Teeth with DD Type I have such poor crown to root ratios that prosthetic replacement including dentures, overdentures, partial dentures, and/or dental implants are the only practical courses for dental rehabilitation. Teeth with DD Type II that are of normal shape, size, and support can be restored with full coverage restorations if necessary. For esthetics, discolored anterior teeth can be improved with resin bonding, veneering, or full coverage esthetic restorations.

Clinicians should be aware that even shallow occlusal restorations may result in pulpal necrosis due to the pulpal vascular channels that extend close to the dentin-enamel junction. If periapical inflammatory lesions develop, the treatment plan is guided by the root length.

Endodontic Considerations

Endodontic therapy, negotiating around pulp stones and through whorls of tubular dentin, has been successful in teeth without extremely short roots. Periapical curettage and retrograde amalgam seals have demonstrated short term success in teeth with short roots.

CLINICAL ALGORITHM(S)

None provided

EVIDENCE SUPPORTING THE RECOMMENDATIONS

TYPE OF EVIDENCE SUPPORTING THE RECOMMENDATIONS

All clinical guidelines are based on 2 sources of evidence: (1) the scientific literature; and (2) experts in the field.

BENEFITS/HARMS OF IMPLEMENTING THE GUIDELINE RECOMMENDATIONS

POTENTIAL BENEFITS

- Reassurance for the patient with heritable dental developmental anomalies and the family
- Reduction in psychosocial consequences of potentially disfiguring conditions
- Prevention of severe attrition associated with enamel loss and rapid wear of poorly mineralized dentin
- Rehabilitation of dentition with severe wear
- Optimization of esthetics
- Prevention of caries and periodontal disease
- Retention of teeth for as long as possible

POTENTIAL HARMS

Not stated

CONTRAINDICATIONS

CONTRAINDICATIONS

The clinician must be cautious in treating individuals with osteogenesis imperfecta (OI) if performing surgical procedures or other treatment that could transmit forces to the jaws, increasing the risk of bone fracture. Some types of protective stabilization may be contraindicated in the patients with OI.

IMPLEMENTATION OF THE GUIDELINE

DESCRIPTION OF IMPLEMENTATION STRATEGY

An implementation strategy was not provided.

IMPLEMENTATION TOOLS

Chart Documentation/Checklists/Forms
Resources

For information about [availability](#), see the "Availability of Companion Documents" and "Patient Resources" fields below.

INSTITUTE OF MEDICINE (IOM) NATIONAL HEALTHCARE QUALITY REPORT CATEGORIES

IOM CARE NEED

Getting Better
Staying Healthy

IOM DOMAIN

Effectiveness
Patient-centeredness

IDENTIFYING INFORMATION AND AVAILABILITY

BIBLIOGRAPHIC SOURCE(S)

American Academy of Pediatric Dentistry Council on Clinical Affairs. Guideline on oral health care/dental management of heritable dental development anomalies. *Pediatr Dent* 2008-2009;30(7 Suppl):196-201. [52 references] [PubMed](#)

ADAPTATION

Not applicable: The guideline was not adapted from another source.

DATE RELEASED

2008

GUIDELINE DEVELOPER(S)

American Academy of Pediatric Dentistry - Professional Association

SOURCE(S) OF FUNDING

American Academy of Pediatric Dentistry

GUIDELINE COMMITTEE

Council on Clinical Affairs

COMPOSITION OF GROUP THAT AUTHORED THE GUIDELINE

Not stated

FINANCIAL DISCLOSURES/CONFLICTS OF INTEREST

Council members and consultants derive no financial compensation from the American Academy of Pediatric Dentistry (AAPD) for their participation and are asked to disclose potential conflicts of interest.

GUIDELINE STATUS

This is the current release of the guideline.

GUIDELINE AVAILABILITY

Electronic copies: Available from the [American Academy of Pediatric Dentistry Web site](#).

Print copies: Available from the American Academy of Pediatric Dentistry, 211 East Chicago Avenue, Suite 700, Chicago, Illinois 60611.

AVAILABILITY OF COMPANION DOCUMENTS

Information about the American Academy of Pediatric Dentistry (AAPD) mission and guideline development process is available on the [AAPD Web site](#).

The following implementation tools are available for download from the AAPD Web site:

- [Dental growth and development chart](#)
- [American Academy of Pediatric Dentistry Caries-Risk Assessment Tool \(CAT\)](#)

PATIENT RESOURCES

None available

NGC STATUS

This NGC summary was completed by ECRI Institute on June 10, 2009. The information was verified by the guideline developer on July 14, 2009.

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